

## General

### Title

Amyotrophic lateral sclerosis (ALS): percentage of patients diagnosed with ALS who were screened at least every 3 months for dysphagia, weight loss or impaired nutrition and the result(s) of the screening(s) was documented in the medical record.

### Source(s)

American Academy of Neurology (AAN). Amyotrophic lateral sclerosis performance measurement set. St. Paul (MN): American Academy of Neurology (AAN); 2012 Jul 31. 75 p.

## Measure Domain

### Primary Measure Domain

Clinical Quality Measures: Process

### Secondary Measure Domain

Does not apply to this measure

## Brief Abstract

### Description

This measure is used to assess the percentage of patients diagnosed with amyotrophic lateral sclerosis (ALS) who were screened at least every 3 months for dysphagia, weight loss or impaired nutrition and the result(s) of the screening(s) was documented in the medical record.

### Rationale

Weight loss is a key prognostic indicator for amyotrophic lateral sclerosis (ALS) with the risk of death increased 7-fold when body mass index is less than 18.5 kg/m<sup>2</sup>. Criteria for gastrostomy are loss of greater than 10% of usual bodyweight since the onset of disease (greater than 5% loss at diagnosis and greater than 5% loss from usual body weight at follow-up). Weight loss is related to bulbar involvement, upper limb disability, depression and/or hypermetabolism. Nutritional status should be checked at 3 month intervals by measuring weight, assessing frequency/severity of choking, duration of meals and

caloric intake (Marin et al., 2011; Lehericey et al., 2012; Spataro et al., 2011; Desport et al., 1999; Vaisman et al., 2009; Dupuis et al., 2008). A speech language pathologist (SLP) helps manage dysphagia to lower the risk of aspiration and optimize oral intake (Kasarskis et al., 1999). Nutritional interventions before deployment of gastrostomy tube may include modification of the texture and consistency of food, and increased caloric intake. Hyperlipemia may significantly prolong survival in ALS but the value of increasing lipid intake is unknown (Marin et al., 2011; Lehericey et al., 2012; Spataro et al., 2011; Desport et al., 1999; Vaisman et al., 2009; Dupuis et al., 2008).

In 9 studies, a total of 469 patients with ALS received enteral nutrition via percutaneous endoscopic gastrostomy (PEG) (Martin et al., 1994; Chiò et al., 1998; Chiò et al., 1999; Del Piano et al., 1999; Desport et al., 2005; Desport et al., 2000; Mazzini et al., 1995; Mitsumoto et al., 2003; Rozier et al., 1991). Using patients as their own controls, 7 studies demonstrated either weight stabilization or modest weight gain over 2 to 24 months (Martin et al., 1994; Chio et al., 1998; Chio et al., 1999; Desport et al., 2005; Desport et al., 2000; Mitsumoto et al., 2003; Rozier et al., 1991). In 2 studies (Del Piano et al., 1999; Mazzini et al., 1995) in which PEG refusers served as controls, weight stabilization was demonstrated in the PEG group vs. continued weight loss in controls ( $p$  less than or equal to 0.03). Enteral nutrition administered via PEG is probably effective in stabilizing body weight/body mass index (Del Piano et al., 1999; Mazzini et al., 1995).

The following clinical recommendation statements are quoted verbatim from the referenced clinical guidelines and represent the evidence base for the measure:

Bulbar dysfunction and nutritional status, including weight, should be checked at each visit (Andersen et al., 2005).

The patient and spouse should be referred to a dietician as soon as dysphagia appears. A speech and language therapist (SLT) can give valuable advice on swallowing techniques (Andersen et al., 2005).

## Evidence for Rationale

American Academy of Neurology (AAN). Amyotrophic lateral sclerosis performance measurement set. St. Paul (MN): American Academy of Neurology (AAN); 2012 Jul 31. 75 p.

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## Primary Health Components

Amyotrophic lateral sclerosis (ALS); screening; dysphagia; weight loss; impaired nutrition

## Denominator Description

All patients with a diagnosis of amyotrophic lateral sclerosis (ALS) (see the related "Denominator Inclusions/Exclusions" field)

## Numerator Description

Patients who were screened at least every 3 months for dysphagia, weight loss or impaired nutrition and the result(s) of the screening(s) was documented in the medical record (see the related "Numerator Inclusions/Exclusions" field)

## Evidence Supporting the Measure

### Type of Evidence Supporting the Criterion of Quality for the Measure

A clinical practice guideline or other peer-reviewed synthesis of the clinical research evidence

A formal consensus procedure, involving experts in relevant clinical, methodological, public health and organizational sciences

A systematic review of the clinical research literature (e.g., Cochrane Review)

One or more research studies published in a National Library of Medicine (NLM) indexed, peer-reviewed journal

### Additional Information Supporting Need for the Measure

Importance of Topic

*Prevalence and Incidence*

Amyotrophic lateral sclerosis (ALS), also known as Lou Gehrig's disease, is a type of motor neuron disease that is a rapidly progressive and fatal neurological disease (National Institute of Neurological Disorders and Stroke [NINDS], 2013).

Twenty thousand to 30,000 people in the United States (U.S.) have ALS (NINDS, 2013).

Five thousand people are diagnosed with ALS in the U.S. annually (NINDS, 2013).

ALS is one of the most common neuromuscular diseases worldwide (NINDS, 2013).

In 90% to 95% of all ALS cases the disease occurs apparently at random with no clearly associated risk factors (NINDS, 2013).

Five percent to 10% of all ALS cases are inherited (NINDS, 2013).

Twenty percent of all familial cases result from a specific genetic defect that leads to mutation of the enzyme known as superoxide dismutase 1 (SOD1) (NINDS, 2013).

No cure exists for ALS. Newer pharmacotherapy agents have been found to reduce the progression, but not halt the disease development (NINDS, 2013).

The prevalence of ALS is said to be between six and eight cases per 100,000 in the population.

Using the higher prevalence estimate and data from the 2000 U.S. census, nearly 22,600 Americans are living with ALS at any one time. Since ALS is a disease of aging, as the U.S. population increases and ages, an increase in the prevalence of ALS can be anticipated (ALS Association, 2012)

Cognitive dysfunction is seen in 20% to 50%, while only 3% to 5% develop dementia that is usually of frontotemporal type (Strong et al., 2009). Consensus criteria for diagnosis have recently been reported (Strong et al., 2009).

Death due to respiratory failure follows on average 2 to 4 years after onset, but a small group may survive for a decade or more (Haverkamp, Appel, & Appel, 1995).

The mean age of onset is 47 to 52 years in familial cases (FALS) and 58 to 63 years in sporadic (SALS) cases (Bobowick & Brody, 1973).

The lifetime risk for developing ALS for individuals aged 18 years has been estimated to be 1 in 350 for men and 1 in 420 for women (Armon, 2007) with male sex, increasing age and hereditary disposition being the main risk factors (Heffernan et al., 2006).

Most patients with ALS die within 2 to 5 years of onset (Lechtzin et al., 2002). Only 10% of ALS patients survive for 10 years or more (Miller et al., "Drug, nutritional," 2009).

Treatment of respiratory insufficiency improves survival, quality of life and respiratory symptoms (Lechtzin et al., 2002; Miller et al., "Drug, nutritional," 2009). The diagnosis and management of respiratory insufficiency is critical because most deaths from ALS are due to respiratory failure (Lechtzin et al., 2002; Miller et al., "Drug, nutritional," 2009; EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis et al., 2012; Laird et al., 2001).

Falls surveillance will lead to interventions to prevent falls and decrease fall related deaths in ALS patients. Falls are an independent predictor of adverse health outcomes (Gil et al., 2008). Fall related deaths occur in 1.7% of ALS patients (Rubenstein & Josephson, 2002). Several specific risk factors for falls have been identified, including muscle weakness, deficits in gait or balance, visual deficits, arthritis, impairments in activities of daily living, depression, and cognitive impairment (Ringholz et al., 2005).

Studies confirm the presence of cognitive impairment in 50% of patients with ALS and particularly implicate executive dysfunction and mild memory decline in the disease process (Laird et al., 2001). More severe impairment occurs in a subset of patients with ALS and has features consistent with frontal temporal dementia (FTD) (Phukan, Pender, & Hardiman, 2007; Gordon et al., 2007). Recent studies have demonstrated the feasibility of screening patients in a busy specialized ALS clinic (Flaherty-Craig et al., 2009; Woolley & Katz, 2011), but this is still not routinely practiced. A fuller characterization of the extent of cognitive and behavioral dysfunction in ALS has important implications given that it shortens survival (Elamin et al., 2011), and the burden and stress for carers of patients with FTD is very great. It also has relevance to effective communication, legal issues and end-of-life decision making by patients with motor neuron disease (MND) (Elamin et al., 2011).

Pseudobulbar affect (PBA), excessive laughing or crying, or involuntary emotional expression disorder affects 20% to 50% of patients with ALS, especially in pseudobulbar palsy (McCullagh et al., 1999). Patients are embarrassed and isolated by these symptoms, which in turn greatly diminishes the patients' quality of life.

Sialorrhea, or drooling, is embarrassing, socially isolating, and is associated with aspiration pneumonia. The prevalence is estimated at 50%, and 70% of patients receiving oral medications for treatment reported benefit (Laird et al., 2001; Miller et al., "Multi-disciplinary," 2009)

Fatigue may be a symptom of depression, poor sleep, abnormal muscle activation, immobility, or respiratory dysfunction. Fatigue diminishes quality of life for patients with ALS. Fatigue was a side effect of therapy in 26% of patients taking riluzole vs. 13% taking placebo (Bensimon, Lacomblez, & Meininger, 1994). Asthenia occurred in 18% of patients taking riluzole vs. 12% of patients taking placebo in a larger study (Lacomblez et al., 1996).

The prevalence of depression in ALS ranges from 0% to 44%, although systematic studies suggest 10% in advanced ALS (Laird et al., 2001; Wicks et al., 2007). Depression shortens survival and lowers quality of life for patients with ALS (Phukan, Pender, & Hardiman, 2007). There is consensus among experts that depression should be treated in patients with ALS (Laird et al., 2001); however, there are no controlled studies of benefit or harm.

Insomnia is common in ALS and may be a symptom of early respiratory weakness, underlying anxiety, depression, or pain (Hetta & Jansson, 1997). There is a concern that sedative/hypnotic agents may suppress the respiratory drive in patients with ALS.

Weight loss is a key prognostic indicator for ALS with the risk of death increased 7-fold when body mass index is less than 18.5 kg/m<sup>2</sup> (Marin et al., 2011; Lehericey et al., 2012; Spataro et al., 2011; Desport et al., 1999; Vaisman et al., 2009; Dupuis et al., 2008).

ALS patients have dysarthria in nearly all bulbar onset patients and nearly 40% of ALS patients with spinal onset. More than 95% of ALS patients cannot speak before death and patients who accept gastrostomy tube, non-invasive ventilation or tracheostomy-ventilation have a greater need for augmentative alternative communication as the disease progresses (Ball, Beukelman, & Pattee, "Communication," 2004; Ball, Beukelman, & Pattee, "Acceptance," 2004; Mathy, Yorkston, &

Gutmann, 2000; Beukelman, Fager, & Nordness, 2011).

End of life discussions will improve patient decision making with respect to disease management (NINDS, 2013; ALS Association, 2012; Strong et al., 2009; Haverkamp, Appel, & Appel, 1995; Bobowick & Brody, 1973; Heffernan et al., 2006). Pain in ALS should be treated following accepted guidelines (Oliver et al., 2011; Albert et al., 1999, Mitsumoto et al., 2005; Nolan et al., 2008; Albert et al., 2005; Albert et al., 2009).

### *Office Visits and Hospital Stays*

One study's significant findings were that common morbidities increased over time (pneumonia [38.1% to 47.3%], respiratory failure [26.9% to 35.5%], and nutritional deficiency [43.0% to 56.3%]); the median length of stay dropped from 6 to 4 days; mean hospital charges increased from \$21,574 to \$24,314; the proportion of hospital deaths decreased over time (17.6% to 14.6%), whereas the proportion discharged to home health/hospice care (14.0% to 18.2%) and to long-term care facilities (13.2% to 27.9%) increased. The odds ratio (OR) of death was 5.03 (95% CI: 4.57 to 5.54) for those admitted with respiratory failure, 1.36 (1.24 to 1.50) for those with pneumonia, and 0.84 (0.77 to 0.92) for those with nutritional deficiency. The high OR of death in patients admitted for pneumonia or respiratory failure is likely associated with more advanced disease, whereas the protective effect of admission for nutritional deficiency is consistent with the predominance of bulbar symptoms and admission earlier in the disease. The trends during the 15 years of this administrative data set were for increasing comorbidities and higher utilization of end-of-life care (Dubinsky, Chen, & Lai, 2006).

### *Family Caregiving*

Caregiver burden was correlated to their level of depression and quality of life and, differently from other chronic disorders, increased with the worsening of patients' disability. ALS patients have a good objective perception of their impact on caregivers (Chiò et al., 2005).

Recent studies assessing caregivers' burden in chronic neurologic disorders have found some features shared by caregivers: the perceived burden exceeds the objective measures of patients' impairment, the amount of burden is independent of diagnosis, and the patients' cognitive functioning is an important factor in determining the level of burden (Thommessen et al., 2002).

### *Cost*

ALS is a difficult to diagnose, fatal, progressive degenerative disease with an average survival time of 2 to 5 years. Percutaneous endoscopic gastrostomy (PEG) and bi-level intermittent positive pressure (BIPAP) ventilation may be the major interventions leading to longer survival of patients with ALS. Riluzole has been shown to have modest effects on survival (as opposed to functional) gains and is currently the only drug approved for the treatment of ALS. Mechanical ventilation (via a tracheostomy tube) is expensive, but is widely used in later stage patients with ALS in the U.S. A review of nine cost-effectiveness studies of riluzole found the following: drug costs and survival gains are the major drivers of cost effectiveness; survival gains are estimated from truncated databases with a high degree of uncertainty; more accurate stage-specific utility weights based on patients who agreed to treatment are needed; case incidence-based evaluations should be carried out; cost-effectiveness ratios are insensitive to discount rates; employment and caregiver issues or externalities have been widely ignored; threshold acceptance cost-effectiveness values are ill-defined and evaluations are not generalizable to other countries because of cost and treatment style differences. On account of the high degree of uncertainty pertaining to survival gains and the relatively high costs per life years or quality-adjusted life-years gained, and while acknowledging that not every therapy has to be cost effective (e.g., orphan drugs), it is still inconclusive as to whether or not riluzole can be considered as cost-effective therapy for ALS (Ginsberg & Lowe, 2002).

### *Disparities*

All races and ethnic backgrounds are affected by ALS (NINDS, 2013).

ALS most common in individuals 40 to 60 years old, but younger and older people can develop the

disease (NINDS, 2013).

Men are more likely to develop ALS than women. Studies suggest an overall ratio of about 1.5 men to every woman who develops ALS in Western countries (ALS Association, 2012).

#### Opportunity for Improvement

The prevalence of malnutrition varies between 16% and 55% in ALS patients across several studies (Genton et al., 2011; Marin et al., 2011; Worwood & Leigh, 1998). There is an adjusted 30% increased risk of death for a 5% decrease from usual weight at time of ALS diagnosis (RR 1.30; 95% CI 1.08 to 1.56). During follow-up, there is an adjusted 34% (95% CI 18% to 51%) and 24% (95% CI 13% to 36%) increased risks of death associated with each 5% decrease in usual weight and each unit decrease in usual body mass index (BMI), respectively ( $p$  less than 0.0001). Malnutrition during the course of ALS was related to a shorter survival ( $p$  equals 0.01), and fat mass level was associated with a better outcome (RR 0.90 for each 2.5 kg fat mass increment) (Marin et al., 2011). Thus, many patients lose weight in ALS and survive for a shorter time.

Treatment to stabilize weight and lengthen survival, with nutritional supplements and enteral feeding, is underutilized. Only 19% of patients utilized nutritional supplements and only 16% of patients utilized enteral feeding in one large study (Miller et al., "Outcomes," 2009).

## Evidence for Additional Information Supporting Need for the Measure

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## Extent of Measure Testing

This measure is being made available without any prior testing. The American Academy of Neurology (AAN) recognizes the importance of testing of all of its measures and encourages testing of the amyotrophic lateral sclerosis (ALS) measurement set for feasibility and reliability by organizations or individuals positioned to do so. The AAN welcomes the opportunity to promote the initial testing of these measures and to ensure that any results available from testing are used to refine the measures before implementation.

## Evidence for Extent of Measure Testing

American Academy of Neurology (AAN). Amyotrophic lateral sclerosis performance measurement set. St. Paul (MN): American Academy of Neurology (AAN); 2012 Jul 31. 75 p.

## State of Use of the Measure

### State of Use

Current routine use

### Current Use

not defined yet

## Application of the Measure in its Current Use

### Measurement Setting

Ambulatory/Office-based Care

Home Care

Hospital Outpatient

Skilled Nursing Facilities/Nursing Homes

### Professionals Involved in Delivery of Health Services

not defined yet

### Least Aggregated Level of Services Delivery Addressed

Individual Clinicians or Public Health Professionals

### Statement of Acceptable Minimum Sample Size

Does not apply to this measure

### Target Population Age

Unspecified

### Target Population Gender

Either male or female

# National Strategy for Quality Improvement in Health Care

## National Quality Strategy Aim

Better Care

## National Quality Strategy Priority

Person- and Family-centered Care

Prevention and Treatment of Leading Causes of Mortality

# Institute of Medicine (IOM) National Health Care Quality Report Categories

## IOM Care Need

Living with Illness

## IOM Domain

Effectiveness

Patient-centeredness

# Data Collection for the Measure

## Case Finding Period

Unspecified

## Denominator Sampling Frame

Patients associated with provider

## Denominator (Index) Event or Characteristic

Clinical Condition

## Denominator Time Window

not defined yet

## Denominator Inclusions/Exclusions

## Inclusions

All patients with a diagnosis of amyotrophic lateral sclerosis (ALS)

Note: Refer to the original measure documentation for International Classification of Diseases, Ninth Revision, Clinical Modification (ICD-9-CM) diagnosis codes and Current Procedural Terminology (CPT) Evaluation and Management (E/M) service codes.

## Exclusions

Documentation of a patient reason for not screening for dysphagia, weight loss or impaired nutrition and documenting the result(s) of the screening(s) in the medical record (e.g., patient declines screening)

Documentation of a system reason for not screening for dysphagia, weight loss or impaired nutrition and documenting the result(s) of the screening(s) in the medical record (e.g., equipment not available to complete the screenings; no insurance)

## Exclusions/Exceptions

not defined yet

## Numerator Inclusions/Exclusions

### Inclusions

Patients who were screened at least every 3 months for dysphagia, weight loss or impaired nutrition\* and the result(s) of the screening(s) was documented in the medical record

*\*Impaired Nutrition Includes:* changes in nutritional biomarkers (serum prealbumin, total protein, or hemoglobin) or body mass index

### Exclusions

Unspecified

## Numerator Search Strategy

Fixed time period or point in time

## Data Source

Administrative clinical data

Electronic health/medical record

Paper medical record

## Type of Health State

Does not apply to this measure

## Instruments Used and/or Associated with the Measure

Unspecified

## Computation of the Measure

### Measure Specific Disaggregation

## Measure Specifics Disaggregation

Does not apply to this measure

## Scoring

Rate/Proportion

## Interpretation of Score

Desired value is a higher score

## Allowance for Patient or Population Factors

not defined yet

## Standard of Comparison

not defined yet

## Identifying Information

### Original Title

Measure #7: ALS screening for dysphagia, weight loss or impaired nutrition.

### Measure Collection Name

Amyotrophic Lateral Sclerosis Performance Measurement Set

### Submitter

American Academy of Neurology - Medical Specialty Society

### Developer

American Academy of Neurology - Medical Specialty Society

### Funding Source(s)

Unspecified

## Composition of the Group that Developed the Measure

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*Patient Representative:* Christine Jasch, OTR/L

*Insurance Representatives:* Fredrik Tolin, MD, MBA (Humana)

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## Financial Disclosures/Other Potential Conflicts of Interest

Unspecified

## Adaptation

This measure was not adapted from another source.

## Date of Most Current Version in NQMC

2012 Jul

## Measure Maintenance

Unspecified

## Date of Next Anticipated Revision

Unspecified

## Measure Status

This is the current release of the measure.

## Measure Availability

Source available from the [American Academy of Neurology \(AAN\) Web site](#) .

For more information, contact AAN at 201 Chicago Avenue, Minneapolis, MN 55415; Phone: 800-879-1960; Fax: 612-454-2746; Web site: [www.aan.com](http://www.aan.com) .

## NQMC Status

This NQMC summary was completed by ECRI Institute on March 8, 2016. The information was not verified by the measure developer.

## Copyright Statement

This NQMC summary is based on the original measure, which is subject to the measure developer's copyright restrictions.

## Production

## Source(s)

American Academy of Neurology (AAN). Amyotrophic lateral sclerosis performance measurement set. St. Paul (MN): American Academy of Neurology (AAN); 2012 Jul 31. 75 p.

## Disclaimer

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